Giant Solitary Fibrous Tumor of the Liver, Case Report: Unusual Site, Extravagant Presentation, Aggressive Behavior

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Abstract

Background: Solitary fibrous tumor (SFT) is a rare fibroblastic mesenchymal tumor. It can originate from abdominal soft tissue which is extremely scarce, and liver indeed is super scant site. SFT liver (SFTL) is mostly benign, but it can be malignant and aggressive with metastases and a potential of recurrence. Hypoglycemia is the least reported feature which is resistant to medical management and will be resolved following resection of a SFT of liver.

Case Report: A 67-year-old man presented with episodes of sudden onset loss of consciousness since 3 months before first visit. He was a previously healthy farmer, without any other complaint or past medical history except for episodes of sever hypoglycemia. Laboratory testing revealed hypoglycemia which was temporarily response to glucose infusion, hypoglycemia was refractive, and whole physical examination and laboratory tests were done and there were no abnormal findings. With probability of finding something such as insulinoma or any other tumor that secreted insulin or insulin-like growth factors, a magnetic resonance imaging (MRI) was performed and revealed an enhancing giant round and well-defined lesion in left lobe of live.

Conclusions: We here present the first case of malignant SFTL presenting with hypoglycemia and metastasis to soft tissue as inguinal bulging 2 months after resection of liver mass, but this particular type of tumor needs more evaluation and investigation.

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Introduction

One of the less discussed entities of neoplasms is mesenchymal neoplasms, and solitary fibrous tumor (SFT) is an extremely rare subgroup of mesenchymal neoplasms. Back in the history, SFT was first differentiated from mesothelioma by Klemperer and Rabin in 1931. Many different names (solely hemangiopericytoma) have been used for the description of this neoplasm. SFT can be seen in every anatomic site but the most common site reported is the SFT of the pleura. Despite serosal site, it can originate from abdominal soft tissue which is extremely scarce, and liver indeed is super scant site, and cases reported in literature is < 50 (mostly benign) until the present day and data about SFT's malignant potential, invasive growth and natural history are not well grounded.

When it comes to diagnosis, it is challenging in terms of pre-operative and even post-operative diagnosis. It is important to exclude other differential diagnosis such as hepatocellular carcinoma, GIST, leiomyoma, and sarcoma. There is a 2:1 female-to-male predominance with the ages of affected individuals ranging from 27 to 83 years (1) imaging is helpful, but it cannot distinguish between malignant and benign tumors. Most often, SFT liver (SFTL) is a solitary encapsulated highly vascular tumor showing heterogeneous enhancement in computerized tomography scans. Diagnosis cannot be certainly obtained based on clinical and radiographic features, and histopathological sampling and immunohistochemistry are the main tools to establish a definite diagnosis (2). Given the limited number of cases reported in literature, it is difficult to establish the long-term prognosis for the patient. Progressively local growth is to be expected. The ability of this tumor to grow to large sizes should prompt strong consideration for resection at the time of diagnosis (2).

The symptoms vary from a totally asymptomatic patient to common symptoms such as abdominal pain and cholestasis (due to compression of biliary duct), and most infrequently patient can present with hypoglycemia due to secretion of insulin-like growth factors (3). Surgery is the treatment of choice. Performing resection with a 1 cm margin, free margins should always be achieved, and in cases where a positive margin is found in the post-operative period,

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re-resection is indicated (4). At present, no role exists for adjuvant medical therapy for this disease. The role of chemotherapy and radiotherapy in these tumors is still controversial and is reserved for cases when resection is incomplete, or there are signs of malignancy (5). There is no solid association between histopathologic finding and SFT behavior which make this tumor even more challenging for every physician. Despite adequate local control, estimated recurrence rates for malignant SFTs are around 30% (34-36%). Locoregional recurrent disease has been shown to be more common in patients with malignant histologic features compared with those with typical histology. While small tumors with low mitotic rates are highly unlikely to metastasize, large tumors ≤ 15 cm, which occur in patients ≤ 55 years, with mitotic figures ≤ 4/10 high-power fields require close follow-up and have a high risk of both metastasis and death (6). The small number of patients with a SFT of the liver and its unknown and unpredictable natural behavior creates the need to a careful registration and follow-up of all identified cases. We presented here the first SFTL presenting refractory hypoglycemia with metastasis in soft tissue which is unrivaled in literature.

Case Report

A 67-year-old male presented with episodes of sudden onset loss of consciousness since 3 months before first visit, he was a previously healthy farmer, without any other complaint or past medical history except for episodes of sever hypoglycemia. Laboratory testing revealed hypoglycemia which was temporarily response to glucose infusion, hypoglycemia was refractive, and a whole physical examination and laboratory tests were done and there were no abnormal findings, with probability of finding something such as insulinoma or any other tumor that secret insulin or insulin-like growth factors, a magnetic resonance imaging (MRI) imaging was performed and revealed an enhancing giant round and well-defined lesion in left lobe of liver, with dimension of 10 * 10 cm (Figures 1 and 2), pancreas was normal.

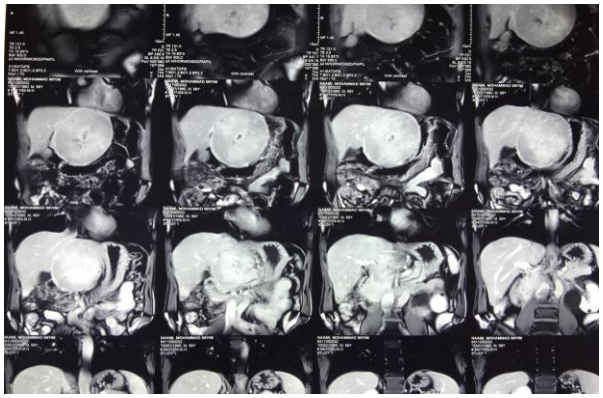

Figure 1. Tumor magnetic resonance imaging originating from left lobe of liver with no local aggression

The other parts of abdomen and pelvis and chest were normal. The patient underwent surgery under general anesthesia and laparotomy was done, in OR, an upper midline laparotomy has provided an excellent access to the abdomen. The abdomen was clean, no sign of ascites and no peritoneal lesion.

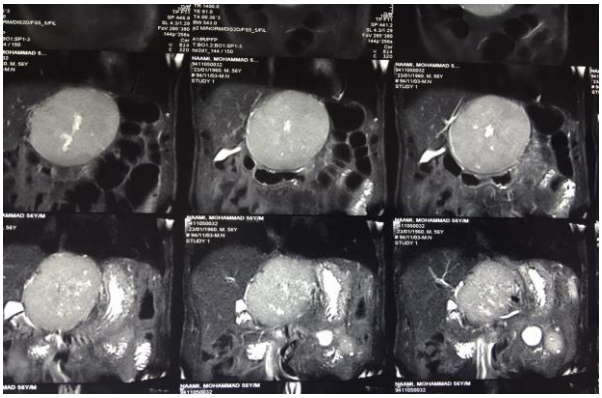

Figure 2. Tumor magnetic resonance imaging -originating from left lobe of liver

As expected, a huge round tumor originating from the left lobe of liver was found with a well-defined capsule and without invasion to the adjacent organs (Figure 3). The frozen section biopsy was done, and the report was a subgroup of sarcoma, left lobectomy of liver was done without subsequent problem in dissecting the lesion. The blood loss was about 600cc which compensated with transfusion. The hemostasis was achieved. The patient's hypoglycemia was improved after surgery, and he was discharged a few days later. Interestingly, permanent pathology report was compatible with SFT of liver. In subsequent follow-up he was symptom free and had no problem, 8 months after surgery, he came back with a bulging mass in right inguinal hernia, incisional biopsy was done and reported synovial sarcoma and he had undergone another surgery again and the lesion was widely excised and sent for pathology, and the pathology report was SFT again, which probably metastasis of liver lesion. In 6 months follow-up after second surgery, he had no other problem. This presentation has never been reported previously in literature.

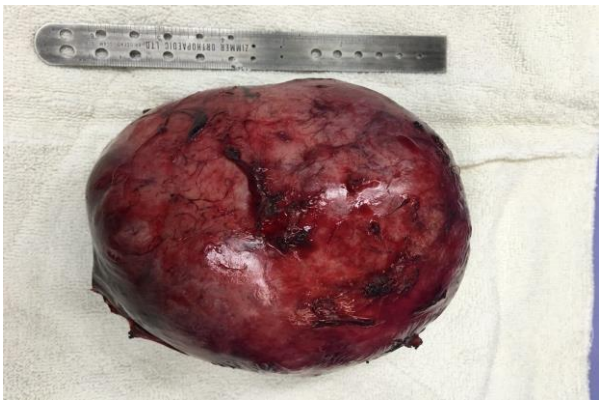

Figure 3. Tumor gross anatomy -note the capsule

Discussion

Evaluating a patient presenting with recurrent hypoglycemia, SFT is the last to come to mind. SFT is a fibroblastic mesenchymal tumor originally described in the pleura (3). Extrathoracic SFTs have been described at almost every anatomic location of human body, but reports of SFT in the abdominal cavity are rare (7). Most extrathoracic SFTs appear to pursue a benign behavior (2). However, SFTL can be aggressive and malignant. Reports of malignant SFTL can be count on the fingers of hands. Most reported site of SFTL is left segment of the liver. Hepatic SFT is very rare, and surgery remains the mainstay of treatment (8). Hypoglycemia is the least reported feature which is resistant to medical management and will be resolved following resection of a SFT of liver and in this case metastases is concurrent with a recurrence of hypoglycemia. SFTs of the liver have a real risk of malignant transformation. The severity of the tumor-associated hypoglycemia may parallel the tumor burden and activity. The syndrome is the systemic effect of IGF2 secreted by the tumor (9). Malignant SFTs involving the liver are currently treated by surgical resection, with the aim of obtaining a margin-negative specimen (10). Taking all together this report of malignant SFTL with only presentation as hypoglycemia, metastasis to soft tissue presenting as a bulging inguinal mass without concurrent hypoglycemia is unique, and he has not been reported previously.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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